TUMOR OF THE FRONTAL SUBCORTEX AND CALLOSUM, WITH FLACCID PARALYSIS OF THE MUSCLES WHICH SUPPORT THE HEAD, APHONIA, MENTAL CHANGE, AND OTHER SYMPTOMS, ILLUSTRATED BY A CASE WITH NECROPSY[1]

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The following case is presented as a contribution to our knowledge of frontal, subcortical and callosal tumors:

Well marked general symptoms of brain tumor—inability to hold the head erect—aphonia—paraparesis, unskilful and uncertain movements and late right hemiplegia—exaggerated reflexes on the right side—involuntary evacuations—mental change—hypesthesia to pain and temperature—tumor involving the subcortex and frontal region of the left hemisphere and the callosum.

The patient, J. B., fifty-two years old, a manufacturer, was in good health with the exception of headaches until about two years before coming under observation. He began to suffer with these headaches at irregular intervals, and later had attacks of dizziness, and much more rarely of sickness of the stomach. He kept a diary, entries in which made during the period between January 9, 1905, and April 26, 1906, showed that he suffered not only from headache and dizziness, but also from rarer spells of nausea, and at times he had sensations of numbness or tingling or some form of paresthesia on his right side, especially in his right upper extremity.

On July 6, 1906, twenty-nine days before the operation, he had a severe spell in which he complained of his head and of great dizziness and plunged or fell towards the right side, going down upon the floor, but not becoming absolutely unconscious. After this he became much worse as regards his headache and general feelings of discomfort. Four days later he was taken to the Long Branch Hospital.

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After his admission to the hospital he grew steadily worse although he had at times periods of moderate improvement with recrudescence of his old symptoms with additional severity. It was noted at the hospital that he tended at times to give way on the right or to fall in this direction, and that he stood and walked unsteadily as if both legs were somewhat weak.

When first examined by me at the hospital he was in bed, where he had been most of the time for some days, although he was taken out of bed daily and put on a chair. His evacuations were usually involuntary. His appearance of face was that of distress and confusion. He understood what was said to him when the matter presented was not too extended or complicated. He answered correctly various questions which were put to him, giving the names in succession of two sons when asked who they were, and also responding in other ways. His responses were always by "yes" or "no" or by one or two words only. He did not talk spontaneously and he did not appear to be able to form sentences of any length. The interne stated that a few days previously to the examination by me he had answered questions, using some short sentences correctly. His speech was not paretic or ataxic, but the little he said was spoken in a very low voice, or a whisper.

Examination of the eyes showed no paralysis of single or associated ocular movements, no ptosis, no difference in the pupils, which were of equal size and responded to light and to converging and diverging movements.

Examination for movements of the jaws and face showed no loss of power in the distribution of the fifth nerve, but drooping of the face on the right and slight impairment of volitional movements on this side. Both eyes could, however, be opened and shut. The patient could not be made to put out his tongue, although he moved it around inside of his mouth.

The right arm was paretic, the impairment of power seemingly being equally distributed throughout the extremity. The patient could, however, at this time use the hand and arm to some extent, as when stimulated to return a grip he grasped the hand of the examiner feebly. The left upper extremity was used with comparative freedom and force, both in response to grasp and other methods of examination, and also spontaneously. The left hand was frequently used to brush away flies, or the hand and arm were moved about in the restlessness and suffering of the patient.

The right lower extremity was distinctly paretic, the impairment of power here being also general, and was, on the whole, about as marked as in the upper extremity. He used the left leg freely but not with full vigor, moving it up and down restlessly and also drawing and pushing it downwards in response to efforts to call forth its use.

All the deep reflexes in the upper extremity of the right side were exaggerated as tested for the elbow and wrist jerks, and also for the knee jerks and the muscle jerks of the lower extremity. Foot clonus was marked and very persistent on the right. The Babinski sign was present, but elicited with difficulty, the plantar reflex response being so vigorous as to confuse the tarsal and the metatarso-phalangeal reflex. The Oppenheim and Gordon signs could be elicited.

Examining for touch, pain and temperature, the patient was moderately hypesthetic to pain and temperature in the right lower and upper extremities. Nothing could be made out as to touch. On account of the patient's mental condition and speech nothing as to topognosis or the senses of position and movement and stereognostic conception could be determined. All the reflexes and sensation were about normal in the left half of the body.

An ophthalmoscopic examination had been made by Dr. William K. Campbell, of Long Branch, who reported that it was difficult, owing to inability to get the patient to fix his eyes, to examine the discs carefully, but that the appearance showed some blurring but not a true optic neuritis.

It was noted that the head of the patient, who was held up and sat on the side of the bed during part of the examination, fell backwards, giving the appearance of a spasmodic retraction of the head. When lying down the appearance presented was also somewhat like this. Examination, however, showed that there was no spasticity of the muscles of the neck and that the head could be readily moved backwards and forwards and to either side by the hands of the examiner. After being thus moved it would again fall backward, the patient apparently not being able to hold it up. The tendency of the head to fall backward seemed to be due to loss of power in the muscles concerned in poising the head and in forward movements of the head.

An effort was made to determine whether or not hemianopsia existed, but without success.

The patient was reëxamined on Monday, July 30, 1906, by the writer, in consultation with the physicians and surgeons to the hospital, and Dr. T. H. Weisenburg. The examination revealed the conditions previously determined, and above noted, with some modifications and additions. In the first place, the paralysis of the leg, arm and face of the right side, especially of the leg and arm, was decidedly more profound than at the examination of three days previous. The patient could not be induced to make any movement of these limbs, nor could he be made to grasp the hand of the examiner. The tests for sensation were repeated with the result of making it more positive that hypesthesia to pain and temperature was present on the right. The observations as to ocular movements and the reflexes were practically the same.

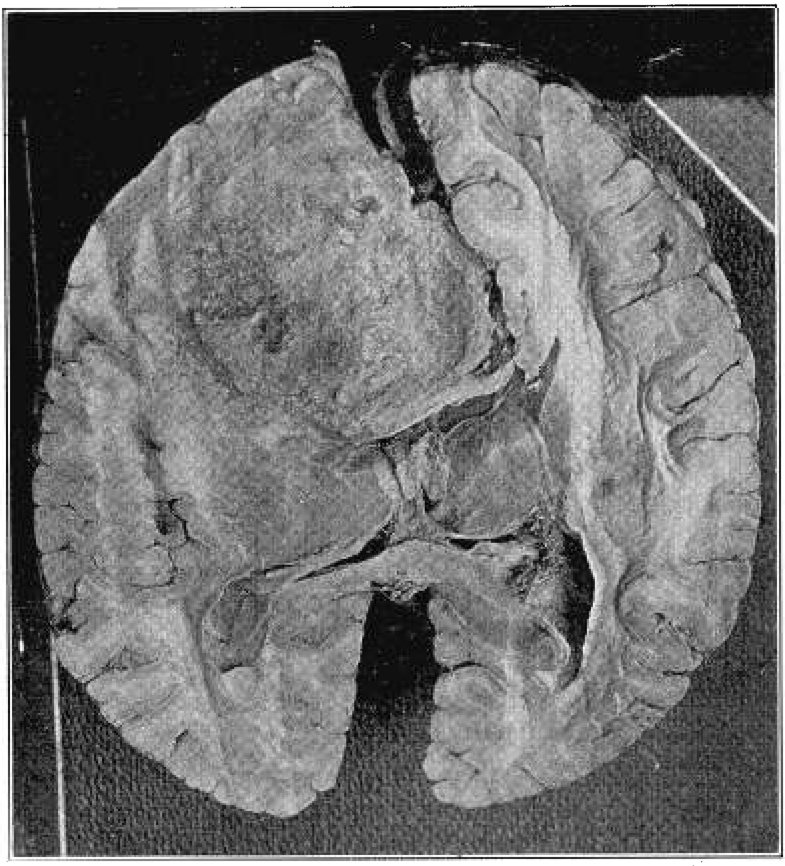
An ophthalmoscopic examination was made by both Dr. Campbell and Dr. Weisenburg. True choking of the discs was not present, but the discs showed blurring of their margins.

On one occasion, about a week before the examination, the patient had complained to his son about hearing some noises, asking the son whether he did not hear them. He apparently indicated the left ear by putting his hand to his ear, but as the right extremity was paretic this use of the left arm may have been due to impairment of power in the right. It was not quite clear whether or not these were real hallucinations.

As the result of the consultations it was decided to operate for decompression and exploratory purposes. Dr. Forman made a large osteoplastic flap, using the DeVilbus rongeur forceps, the opening being so planned as to be about two thirds in front and one third behind the central fissure. Its superior line was three and one fourth inches in length. The sides of the opening, about three inches in length, trended to a base line of about one and one half to two inches. Before and after the exposure of the dura the brain showed great tension as if a subcortical tumor might be present, but no growth was revealed by the exploration and it was deemed best to close the opening. The patient died within twenty-four hours after the surgical procedure.

At the necropsy which was performed by Dr. Weisenburg, the dura was somewhat adherent to the brain at the site of the operation. The superior, middle and inferior frontal convolutions were flattened and the fissures between them were almost obliterated. The orbital surface did not show any compression. Longitudinal sections of the brain made at various levels showed a large tumor which involved the interior of the left frontal lobe, compressing the right hemisphere as well as the left in a manner to be described. The tumor was soft in consistence, gelatinous in its structure, its border infiltrating into the surrounding brain tissue. It involved nearly the whole of the subcortex of the left frontal lobe. In its superior portion it was within one fourth of an inch of the cortex; anteriorly it was within one eighth to one fourth of an inch of the frontal tip; inferiorly it was within one half of an inch of the orbital surface, and laterally to within one half of an inch of the cortex. In the median portion in its widest area it projected to the right side, compressing the right frontal lobe, including the subcortex directly underneath the precentral convolution. Posteriorly the tumor compressed on the right side the caudate nucleus and to some extent the anterior limb of the internal capsule, but did not press upon the knee and posterior limb. It apparently here had made pressure upon some of the motor fibers coming from the precentral convolution. On the left side the tumor had compressed out of their proportionate relations the left caudate nucleus, the anterior limb of the internal capsule, the lenticula and the anterior portion of the thalamus.

The anterior limb of the internal capsule formed with the posterior limb a right angle and the posterior limb was considerably distorted. The tumor had obliterated the anterior horns of the lateral ventricles on both sides and extended as far back as the anterior portion of the third ventricle which was not compressed. The tumor had destroyed entirely the anterior and middle portions of the callosum. In its widest portion it measured three and one fourth inches in width and two and one half inches antero-posteriorly, although it was probable that it extended further by infiltration than was shown by the macroscopic examination. It was a sarcoma.



Photograph of a section through the upper part of the basal ganglia.

Sections made of the pons and peduncle showed a slight secondary degeneration of the pyramidal fibers of the left side.

The patient in this case had, in the first place, some of the well known symptoms of brain tumor, such as are usually present in a lesion of this description wherever situated. These were pain in the head, vertigo or dizziness, and a blurring of the optic discs which probably represented the beginning or the imminence of optic neuritis.

Among the most striking symptoms which seemed to be focal in character were the awkwardness or clumsiness of movement in the extremities of the right side of the body, which towards the termination of the case took the form of a hemiplegia; the weakness or paraparesis of both lower extremities, which was manifest before the right-sided hemiplegia became apparent; the aphonia or lowering of the voice; the tendency of the head to fall backwards, which was apparently due to a paresis or paralysis of some of the muscles concerned in holding the head erect, no spasticity being present; the involuntary evacuations from both bladder and bowels; and the mental apathy or feebleness. The deep reflexes in the right half of the body were exaggerated, and some hypesthesia to pain and temperature was present.

These symptoms are to be attributed in the first place to the large amount of destruction of the subcortex of the frontal lobe, and in the second place to the destruction of the knee and the anterior half of the callosum.

The hypesthesia to pain and temperature is probably to be referred to the involvement directly or by pressure of the subcortex of the gyrus fornicatus.

The exaggeration of the deep reflexes on the right side, including the Babinski sign and foot clonus, which were probably late symptoms, had for their pathological cause compression or direct implication of the pyramidal tract, the posterior extremity of the tumor having encroached somewhat upon the motor subcortex and having produced distortion of this portion of the brain and of the posterior limb of the internal capsule.

Such mental apathy, weakness, and disturbance as were present may be referred jointly to the involvement of the frontal lobe and the callosum. It is well known that psychic symptoms have often been observed in frontal and especially pre-frontal tumors, this being generally more marked when the neoplasm is situated in the left hemicerebrum. Mental symptoms have also been reported in some cases of callosal tumor.

The involuntary evacuations which were present for some days before the death of the patient furnished an interesting feature of the disease. In many cases of tumor of the brain the patients retain control over the bladder and bowels to a very late period, although this is not retained in other cases. The question of the position of the centers controlling these movements is interesting. Anal, vaginal and vesical centers have been placed in the paracentral lobule. The sphincters of the anus, bladder and vagina are doubtless controlled by cortical centers, the centers of one side being sufficient for this control. Destruction of the cortical centers of one side would not cause total paralysis of the sphincter muscles of the anus, bladder and vagina, whereas if these were destroyed on both sides this result would follow; and similarly it would be expected that destruction of the fibers going from these centers, or the commissural fibers connecting these centers through the callosum, would bring about this result.

To the writer the most interesting focal manifestation in this case was the tendency of the head to fall backwards, apparently because of paralysis or paresis of the muscles which hold the head in the erect position. This form of paralytic affection has, so far as I know, not been recorded. In another case recently observed and placed on record at the same meeting at which the present case was reported, a paralysis or extreme hypotonia of the muscles which support and poise the head was the feature of most interest, although the case in other particulars differed from that here presented and was probably an instance of cerebellar disease. In both of these cases no spasticity was present, but the reverse. The patient's head tended constantly to fall backwards, and apparently could not be supported in an erect position. Even when lying down this tendency was apparent and led to an examination being made for rigidity of the neck muscles, which however was absent; the head was easily movable in all directions by the hands of the observer, offering absolutely no resistance. It was clearly not a case of spasmodic retraction.

Whether the destruction of the callosal commissural fibers between the cerebral regions for the movements of the head and neck may not have had something to do with this symptom, is worthy of consideration. The head is carried erect and is poised in various positions through the interaction of a complicated bilateral musculature, all parts of which are probably governed largely by centers in each hemisphere. Destruction of these centers on one side produces therefore no appreciable effect; destruction of them or of their subcortical connections on both sides might produce a marked bilateral paralytic effect, and by the same reasoning, destruction of the callosal commissural tracts might produce inability to use these muscles.

Mott and Schäfer, as cited by the writer in his book on "The Nervous System and Its Diseases," "experimented with weak induction currents on the callosums of monkeys, and produced localized bilateral movements in all parts of the body. From before backward, in order, in different segments of the callosum, electrical stimulation caused movements of the head and eyes, the face, the shoulder, the trunk muscles, and the legs and tail. Cutting away one hemicerebrum, they found that by stimulating a thin middle strip of the callosum localized movements were produced on the side of the section, that is, on the side of the body opposite to that hemicerebrum with which the callosum was still connected, the order of movements being the same as above mentioned." As the result of these experiments, it would seem that a large destructive lesion on one side of the cerebrum, involving the anterior part of the callosum and the adjoining portion of the other hemisphere, would cause more or less bilateral paralysis of the parts controlled by the centers whose commissural connections were destroyed. These connections in the case here recorded were probably those of the centers concerned with movements of the head. These centers, it is well known, are situated well forward in the motor region, in such position that the tumor found in this case would be likely to destroy both their projection and commissural tracts.

The aphonia or apparent aphonia may have been merely a late symptom indicating great weakness, or may have been dependent somewhat on destruction of the commissural connections between the laryngeal centers. The patient also had apparently some aphasia, but this aphasia seemed to be more than it really was because of his loss or great impairment of vocalizing power. One, two and three days before death he responded clearly to questions asked him by "yes" or "no," or made use of single words, or even short sentences, but it was always necessary to have the ear of the listener close to the face of the patient in order to understand what he said. The following are some illustrations of conversation held in this way: When a watch was held before him, after looking at it intently, he said "watch" in a whisper or very low tone; other objects, as a pencil and key, immediately afterwards presented to him, he called also a watch in the same tone.

Several days before his death the patient became unable to stand or walk without support, this apparently being due to great clumsiness and weakness of the right half of the body, which became a hemiplegia shortly before death, and also perhaps to some extent to the much less degree of weakness of the left side. A species of paraparesis has been observed in some cases of callosal tumor. The hemiparesis or hemiataxia, or clumsiness of the limbs on the right side was doubtless due to the prefrontal and midfrontal disease, the late hemiplegia resulting from pressure and direct encroachment on the motor region.

I have reported this case as one of somewhat unusual character in that it presented at least two symptoms unique or almost unique, as well as other well-known manifestations of frontal, subcortical, and callosal tumors. The literature of callosal disease has recently been well presented by Bruns, who, including his own, collected twenty-six cases in which this structure was involved. In referring the clinical phenomena to the lesion found, most stress should of course be laid upon the frontal subcortical growth, this being very large, although the involvement of the callosum must also have recognition.

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